

Distal renal tubular acidosis presenting with hypokalaemic paralysis in pregnancy.

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Case report

Presentation

- 31year P⁰⁺¹ 32⁺⁵ brought to maternity triage by ambulance with generalized weakness after vomiting and diarrhea for 1 week.
- She had MRC grade 1/5 power in all four limbs with normal sensation, reflexes and plantar responses. Blood tests revealed a potassium level of 1.8mmol/L and a bicarbonate level of 9mmol/L.

Background

- BMI 41
- Bipolar affective disorder on Pregabalin, Mirtazapine, quetiapine.
- Family history of Postpartum psychosis
- Previous alcohol abuse – attends AA
- Previous cocaine abuse attends NA
- Current smoker
- Previous Bulimia

Investigations

- FBC, CRP, U&E, LFT, bone profile, amylase, lactate, ABG, MSU, stool culture
- ECG
- BBV screen, vasculitis screen, liver screen
- Pituitary, adrenal work up
- Protein electrophoresis
- Urine, serum osmolality
- USS abdomen, CT hear, MRI pituitary

Day 1 –
Triage

MEOWS 0
MRC 1/5 power, Potassium 1.8, HCO₃ 9, ECG ST depression
LW MDT

Day 1 –
CCU

Potassium replacement with cardiac monitoring
Neurology/ Gen Medicine input

Day 3–
CCU

Slow rise in K⁺
Hypertension, Liver derangement
Liver screen

Day 4–
Labour
Ward

Coagulopathy with normal platelets - Provisional diagnosis AFLP
Antenatal steroids
Caesarean Section

Day 5 –
Labour
Ward

Encephalopathy
Diabetes Insipidus
Gastro/ Endocrine/ Renal

Day 6 –
Labour
Ward

Desmopressin, Hydrocortisone
SLTU input

Day 7–
Medical
HDU

Hypokalemia, normal anion metabolic acidosis, encephalopathy
– dRTA

Day 11 –
16

Transferred to postnatal ward and discharge

6 week

Postnatal Debrief
Neonate suspected to have fatty acid oxidation disorder -genetic
screening awaited

3 months
Renal F/u

Continuing HCO₃ and Potassium replacements

Discussion

Distal Renal Tubular Acidosis

- Renal tubal acidosis is a disorder of acid base regulation in the kidneys, leading to normal anion gap metabolic acidosis.
- Distal renal tubular acidosis is typified by the presence of hypokalaemia, metabolic acidosis and an elevated urine pH¹It can be caused by genetic mutations, autoimmune conditions (e.g. Sjogren’s syndrome) and medications (e.g. lithium).
- Physiologic changes in pregnancy may worsen pre-existing renal tubular acidosis.
 - mild respiratory alkalosis with some urinary bicarbonate loss as a compensation.
 - The volume of distribution for bicarbonate increases during pregnancy.
- Its effects on pregnancy and vice versa are not known, but chronic acidosis may affect fetal bone growth and development.
- Chronic metabolic acidosis can compromise fetal circulation and lead to fetal distress and demise.²
- Treatment includes replacement of potassium and bicarbonate.

Acute Fatty Liver of Pregnancy

- Rare 1 in 7000 – 1 in 20,000
- Common in Primigravida, male fetus, multiple pregnancy
- UKOSS 2% maternal mortality, 11% perinatal mortality
- Severe vomiting 60%, Abdominal pain 60%, 3-10 folds transaminitis, jaundice within 2 weeks, coagulopathy 90%, profound hypoglycemia 70%, renal impairment, diabetes insipidus.
- Diabetes insipidus presents with excessive thirst and polyuria due to relative deficiency of vasopressin (ADH).
- Transient diabetes insipidus due to decreased vasopressinase breakdown by the liver is found in association with AFLP.
- Coagulopathy in the absence of thrombocytopenia differentiates AFLP from HELLP syndrome.
- Expediting delivery of the fetus, multidisciplinary input and aggressive management of hypoglycaemia and coagulopathy are associated with improved prognosis in AFLP.^{3,4}

Conclusion

Medical disorders in pregnancy often impose a challenge to the Obstetric team. Multidisciplinary involvement and appropriate timing of delivery leads to improved outcomes.

References

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